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Spindle Cell Fibromatosis of Right Arm

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Spindle cells named on the basis of their appearance under microscopy are found in both healthy tissues and tumors. Spindle cells look long and slender shaped cells on microscopy. Extra abdominal Fibromatosis is common in patients between puberty and 40 years of age group with female predominance. A 15 years old male admitted with complain of pain and swelling in his right arm for 1.5 years without any history of antecedent trauma. Swelling was insidious in onset and gradually progressed to become the size of about 8 *6 cm.

MRI of right arm showed lobulated altered signal intensity lesion of size 63*41*84mm (AP*TR*CC) in lateral aspect of mid arm involving deltoid and triceps muscle with few enlarged lymph nodes. PET CT was suggestive of FDG avid soft tissue density in the lateral aspect of right mid arm involving mid arm muscles sparing the underlying bone and overlying skin with ipsilateral axillary lymph nodes involvement.

Using posterior lateral approach to arm wide excision of the tumor done protecting the radial nerve. Postoperative follow-ups showed no neurological deficit in upper limb.

Keywords: Spindle cells; fibromatosis; extra abdominal fibromatosis; tumor; high grade malignant tumors; neurovascular deficit.

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1. INTRODUCTION

Spindle cells named on the basis of their appearance under microscopy are found in both healthy tissues and tumors. Spindle cells look long and slender shaped cells on microscopy[1].

In healthy tissues spindle cells found as a part of fibroblasts, myofibroblasts, neural, epithelial and vascular tissues. Fibromatosis is a benign tumor commonly presents in abdomen and extremities [2].

Extra abdominal Fibromatosis is common in patients between puberty and 40 years of age group with female predominance. Fibromatosis is caused by proliferation of highly differentiated fibroblasts [3,4]. These types of tumor usually do not metastasize but local infiltration and compression to surrounding vital structures is common [5].

2. CASE REPORT

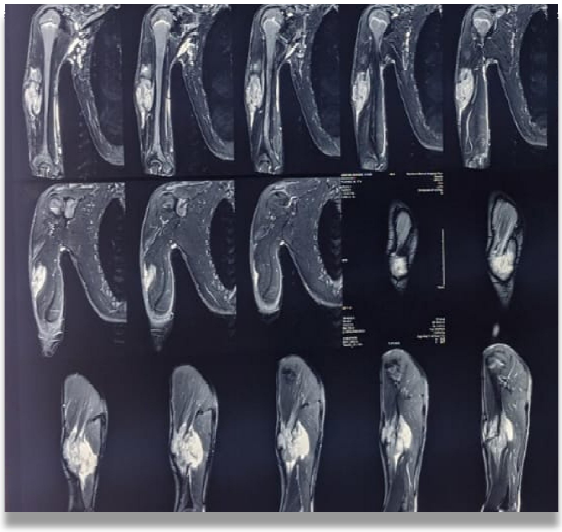
A 15 years old male admitted with complain of pain and swelling in his right arm for 1.5 years without any history of antecedent trauma. Swelling was insidious in onset and gradually progressed to become present size. Patient didn't provide any history of alcohol consumption or smoking. There was no history of any cancer in his family members.

On examination patient appeared to be well built. On examining the local site, there was a swelling of 8*6 cm of size over antero-lateral aspect of right arm. Overlying skin was normal. Swelling had smooth surface, firm consistency and irregular margins were palpable. The swelling was non mobile, non fluctuant and non pulsatile. Local temperature was raised and deep tenderness over swelling was elicited. Few axillary lymph nodes were palpated. No any other swellings were palpated elsewhere in the body. Elbow extension was limited. There were no signs of radial nerve entrapment.

Routine blood investigations such as complete hemogram, liver and kidney function tests, alkaline phosphatise etc. were obtained and found to be within normal ranges. CT of right arm showed large mildly hypo dense soft tissue lesion in anterolateral aspect of right arm involving muscular and inter muscular plane with no oblivious interosseous extension or osseous erosion.

MRI of right arm showed lobulated altered signal intensity lesion of size 63*41*84mm (AP*TR*CC) in lateral aspect of mid arm involving deltoid and triceps muscle with few enlarged axillary lymph nodes. The lesion appeared hypointense lobulated lesion with multiple flow voids within and hyperintense on T2WI/STIR. The MRI features were suggestive of soft tissue neoplasm of mesenchymal origin with differential diagnosis of round cell neoplasm and myositis ossificans.

PET CT was suggestive of FDG avid soft tissue density in the lateral aspect of right mid arm involving mid arm muscles sparing the underlying bone and overlying skin with ipsilateral axillary lymphnode involvement.



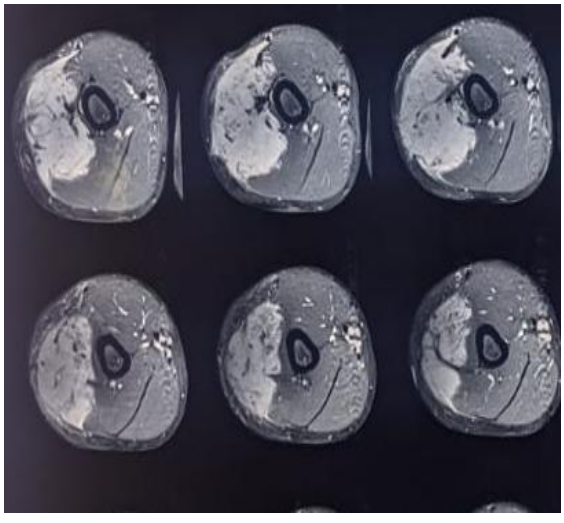


Fig. 1, 2. MRI of right arm

Patient was taken for surgery after obtaining consent regarding possible radial nerve palsy. Using posterior lateral approach to arm wide excision of tumor done protecting the radial nerve. After soft tissue dissection interval made between lateral and the long head of triceps the radial nerve was identified in spiral groove. While tracing the radial nerve distally no entrapment was found. Then the tumor was excised with normal tissue margins confirmed by frozen section biopsy. Tumor along with few pieces from deltoid, triceps and brachioradialis and few pieces of periosteum obtained from humerus shaft sent for histopathological examination.

Histopathological examination was suggestive of spindle cell neoplasm consistent with fibromatosis. Periosteum and all muscle pieces were found to be un-involved by tumor cells. All the margins were tested negative. Tumor cells were S-100 negative, desman negative, SMA positive, CD34 negative with Ki67 -1-2 %.

The patient was kept in post operative ward and discharged after seven days. Patient was followed up regularly for three months. Wound assessment was done; muscles power and sensations were tested. No neuromuscular weakness or paraesthesia was noted.

3. DISCUSSION

Spindle cell neoplasms cover a wide spectrum of diseases ranging from reactive tumor like lesions to high grade malignant tumors [6]. Spindle cell fibromatosis are the benign tumors originating from clonal proliferation of spindle cells. The incidence of these tumors is three in every 3.5 million with a 2:1 female: male predisposition [8]. Possible risk factors include female sex (female: male =2:1), a previous history of trauma/surgery or pregnancy [8,7,9]. In our patient there was no identifiable risk factor.

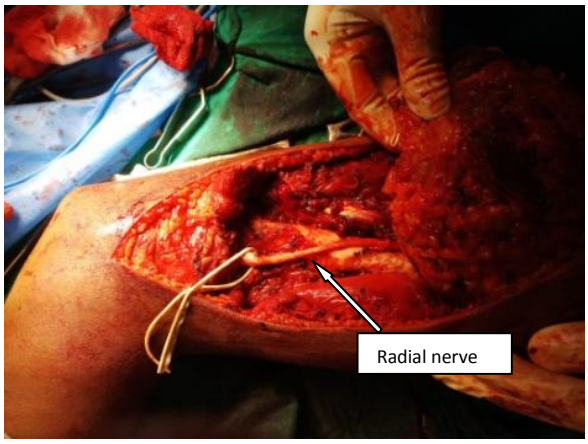


Fig. 3. Intra-operative picture of tumor with intact radial nerve

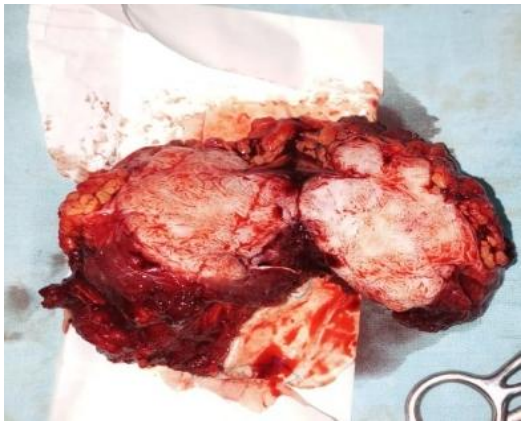


Fig. 4. Specimen of resected tumor

Fig. 5, 6. Postoperative clinical photograph with intact neurology

Considering their rare occurrence the correct and timely diagnosis is crucial for their management. Although some studies suggest that only 50% of the patients are diagnosed correctly [9].

Fibromatoses are known to be benign aggressive tumors without metastasising nature but with extreme high recurrence rate [8,10]. Mostly originating from the deep muscle layers or inter muscular plane they have tendency to infiltrate locally [10]; hence early diagnosis and early intervention is crucial in such cases to prevent further damage to the other vital structures nearby.

Due to their variable clinical presentation and biological behaviour, a multi disciplinary approach with collaboration among medical oncologists, surgeons, pathologists and radiologists is necessary.

Limb salvage surgeries such as wide resection of tumor accompanied with radiotherapy is the preferred treatment in most of the aggressive Fibromatosis tumors [8].Several studies suggest about complete resection with tumor free margin is very crucial. However having positive resection margins may not predict recurrence [11]. About two third of the patients reported recurrence regardless of their treatment modalities [7]. The aggressive nature of the tumor makes the complete resection difficult without sacrificing adjacent vital structures [10].

Our patient was a 15 years young male with swelling and pain in his right arm without any risk factor for extra-abdominal fibromatosis. After radiological and histological investigations surgery was planned. In this case we did wide resection of the tumor with taking negative margins from adjacent tissues and preventing the radial nerve which was prone to compression and infiltration from the tumor. In postoperative follow-up the functional outcomes were satisfactory and there was no neurovascular deficit.

4. CONCLUSION

Fibromatoses of extra abdominal origin are rare and can be mis-diagnosed if not approached properly. Fibromatoses of extremities are rare and raises clinical interest due to their local aggressive and infiltrative nature. The diagnosis should be confirmed only after detailed radiological and histological examinations. Due to their high recurrence and rate complete resection is necessary and the locally aggressive nature of these tumors compels the surgeon to operate as early as possible. We propose that if extra-abdominal fibromatosis is considered as a differential diagnosis in any patient, it should be imaged appropriately (best with MRI) and confirmed by histological examination and should be excised with safe margins as early as possible.

CONSENT AND ETHICAL APPROVAL

Patient was operated after obtaining informed consent and there were no ethical issues.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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